Since Pierre Robin first described the syndrome of hypoplasia of the mandible, cleft palate, glossoptosis, inspiratory retraction of the sternum, cyanosis, and malnutrition which has come to bear his name [2, 3], numerous reports of cases have appeared in the literature. The fatal termination of some cases is a testimony of the serious nature of the syndrome. Although a variety of mechanical and surgical therapeutic procedures have been suggested [4–11], a rationale for the proper management of individual patients has not been adequately presented. It is the purpose of this communication to present observations from the serial studies of patients with this disorder, upon which a physiologic approach to management may be based.

It is generally agreed that the pathophysiologic events in this syndrome are as follows: the receding chin fails to support the tongue in its normal forward relationship and hence fosters the glossoptosis. The retroposed tongue impinges against the posterior wall of the pharynx, obstructing inspiration and impeding feeding. Slight excess in mucus or saliva tend to enhance the pharyngeal obstruction and may precipitate severe cyanotic seizures, resulting in death. Starvation and respiratory infections, or both, may follow as a consequence of the chronic glossoptosis.

Since the major symptoms are definitely related to the micrognathia, it was of special interest to focus our attention upon the growth of the mandible in these infants. In reviewing the published case reports, it appeared that two questions, basic to an understanding of a more appropriate management of these cases, remained unanswered. The first question could be phrased as follows: How soon would mandibular growth be sufficient to accommodate the tongue and hence insure a more adequate airway? This is assuming that the infant could be placed in an adequate metabolic climate. The answer to this question would have an important bearing on immediate management. The second question is related to whether or not mandibular growth would be sufficiently sustained to provide an esthetically satisfactory facial profile.

The answer to these questions had to await the development of accurate roentgenographic techniques for measuring the growth of the head in infants, which made it possible to undertake a serial study of the growth of the micrognathic mandible. The results of these investigations have provided useful information as a basis for therapy and prognosis.

During the past 25 years, cephalometric roentgenography has produced valuable information pertaining to the growth of the head in normal [12] and pathologic conditions [13] in man. In addition to the cephalometric roentgenographic measurements, dental impressions of the maxillary arch were obtained at regular intervals. The impressions were cast in dental stone and then subjected to various measurements. The infants were recalled for study every 3 months during the first year of life and twice annually until the age of 5 years. Thereafter, they were observed annually.

From a large series of similar cases now under longitudinal study at the Cleft Palate Center of the University of Illinois [14] we have selected three cases for presentation. At birth, each of these infants present an isolated cleft of the palate and mandibular micrognathia. Despite these similarities, certain important differences existed to vary the management and progress of each infant. The differences to be described in each of these cases represent the major variations which we have encountered in our experience with this syndrome. For the sake of brevity, the case history will be confined to such information as is directly pertinent to the purpose of this paper.
8.1.1 Case 1

At 2 months of age, D.R.P., a small white dehydrated baby girl, was admitted to the Research and Educational Hospital, with the principal complaint of intermittent pneumonitis for the previous 5 weeks. After a normal full-term spontaneous delivery, it was noted that the baby had an isolated cleft palate and a small mandible. The birth weight was 5 lb. 13 oz. (2,640 g). Feeding had been difficult because of the tendency of the baby’s tongue to fall back into the pharynx. There was no history of a cleft on either side of the family. The mother gave no history of being exposed to a contagious disease or other illness during her first trimester of pregnancy.

Because of the patient’s congenital defects, she presented primarily a feeding problem. An attempt was made to design an obturator so that the baby could be bottle-fed. This was met with failure, and gastric gavage was necessary. However, the patient did not gain weight and continued to do poorly. Approximately 1 month after admission, it was considered that a tracheotomy was the only procedure which might possibly save the baby’s life. After the tracheotomy, respiration was greatly facilitated, and soon the patient was removed from the oxygen tent. Her improvement thereafter was constant. The patient was discharged from the hospital 3 months after admission, at 5 months of age.

Growth Studies: Oral examination disclosed a cleft involving the one half of the hard palate and extending posteriorly throughout the length of the soft palate. Ptosis of the tongue was evident.

On the fifth day following the tracheotomy, at the age of 3 months 1 day, the first lateral film was obtained without sedation. A tracing of this film revealed that the head was in slight dorsiflexion, the position of greatest comfort for the infant. The relative smallness of the mandible and its effect on the facial profile was self-evident. In relation to contiguous anatomical structures, the posture of the tongue was abnormal. It projected through the cleft in the palate into the nasal cavity. (Fig. 8.2)

In the lateral head plate, the dorsum of the tongue was visible at a level above the palatal plane. Posteriorly, the tongue impinged on the airway. The posterior surface of the tongue, just above the epiglottis, was in close approximation to the posterior outline of the pharyngeal wall. At this level, the airway was almost completely occluded. The tracheotomy tube was in place. The airway in its posteroanterior dimensions is fairly wide, and the tongue occupies a more protrusive relationship to the mandible.

The second film in Fig. 8.2, obtained less than 6 weeks later, at the age of 4 months 13 days, revealed considerable enlargement of the airway. At the same time, a considerable increment in the growth of the head and, above all, of the mandible was recorded. Subsequent films, up to the age of 3 years 5 months 12 days, gave proof of the gradual and continued growth of the lower jaw in relation to the total face and the increase in the dimensions of the pharyngeal airway. At 8 months following removal of the tracheotomy tube, the air passage was demonstrated to be quite adequate (Fig. 8.2).

The superimposed tracings, from 3 months 1 day of age to 3 years 5 months 12 days, reveal the progressive

---

Fig. 8.1. a Serial cephalometric tracing of a child with Pierre Robin Sequence showing a severely micrognathic mandible. At 3-4-13 there is a big increase in the pharyngeal space (darkened area). b Superimposed tracings at 2 months and 10 days (0-2-10) and 3 years, 4 months, and 13 days (3-4-13) shows a rapidly growing micrognathic mandible. (Reprinted with permission from [1])
growth of the cranial vault, the maxilla, and the middle face and the increase in length and height of the mandible (Fig. 8.1b). Gradual improvement in the facial profile was recorded by the changes in the facial angle and in the angle of convexity. The facial angle is a measure of the degree of protrusion or recession of the chin. In this instance, the facial angle increased from 64°, at 3 months of age, to 70°, at the age of 3 1/2 years, indicating a reduction by 6° in the recessiveness of the chin. While the mandible is still in a retrusive relation to the rest of the face, the potential for further improvement with continued growth still exists.

The changes in the angle of convexity were more interesting. This measurement relates the maxilla to the total facial profile. At 3 months of age, the angle of convexity was 140°, and at 3 1/2 years it measured at 154°. The integrated growth of the several areas of the face was such as to improve the overall configuration of the facial profile. Serial photographs at 2 months, at 13 months, and at 3 years of age further testify to the changes in this child’s face. In the last photograph, the patient is posed beside her older sibling.

The changes in the position of the hyoid bone are of particular interest, insofar as they reflect a change in the relative position of the tongue. The tongue is composed of several individual muscles originating from the base of the skull, the mandible, the hyoid bone, and the walls of the pharynx. Changes in the position of any of its bony or fibrous attachments would...
tend to reflect on the position of the tongue. Conversely, changes in the posture of the tongue would reflect on the spatial relations of the mandible and hyoid bone. Therefore, to study the position of the hyoid bone is, in a sense, to study the position of the tongue. With growth there occurs a forward and downward migration of the hyoid bone from the base of the skull. The pattern of changes in the posture of the hyoid bone observed in this patient sheds further light on the favorable adjustments consequent to growth. During the first 5 months of our studies, the hyoid bone migrated downward and forward. This resulted in an increase in the angle S-N-H. But, from 8 months onward, this angle became fairly stable and the hyoid bone began to descend principally in a downward direction.

Comment: This case was selected to typify the findings in several similar cases, one of which has been followed to the age of 7 years. Not all cases of Pierre Robin syndrome present such acute histories. When clinical evaluation suggests that there will be no improvement or that possibly death may ensue, tracheotomy should be undertaken without hesitation to prevent further aggravation of the symptoms. Once an adequate respiratory exchange was made possible, improvement in oxygenation and feeding followed. In such instances, we have recorded rapid growth and favorable changes in the facial appearance.

8.1.2 Case 2

J.G, a white girl, was referred to the outpatient clinic of the Cleft Palate Center at the age of 2 months with a diagnosis of cleft palate and mandibular micrognathia. Following an uneventful pregnancy, the delivery was normal and at full term. The birth weight was 6 lb. 11 oz. (3,030 gm). The infant had some difficulty in breathing, but this was relieved by placing her in a prone position. Tube feeding was employed for the first few days after which she was given bottle feedings. At 6 days of age, the infant was discharged from the hospital. There was no family history of cleft palate. The mother suffered no illness during her pregnancy.

Oral examination revealed an unusually small tongue closely attached to the floor of the mouth. In the course of our first examination under sedation, the infant became cyanotic and failed to initiate mandibular movements sufficient to permit the passage of air. This was relieved immediately by maintaining forward traction on the tongue and mandible. After about 5 min, the infant recovered control of mandibular movements, and respiration normally. Aside from this isolated episode, which occurred under sedation, the parents did not report any similar difficulties. The child has continued to grow and develop at a satisfactory rate.

Growth Studies: The casts disclose symmetrical cleft of the hard and soft palate, extending distally from the region of the nasopalatine foramen. Additional casts obtained at regular intervals revealed that the cleft had narrowed, so that it now presents a narrow V-shaped defect (Figs. 8.3, 8.4).

The earliest lateral head palate, at 2 months 10 days of age, displayed a small mandible and small tongue. The latter was positioned high and above the floor of the nose, but relatively remote from the posterior wall of the pharynx. The airway appeared sufficient to sustain respiration without any undue effort on the part of the infant. Progressive growth changes recorded up to the age of 3 years, 4 months, 13 days disclosed mandibular growth and generalized growth in all areas of the face and cranial vault. Mandibular growth was continuous and progressively downward and forward. During the period studied, from 2 to 40 months of age, the facial angle increased from 61.5, becoming more obtuse. The angle of convexity increased from 147° to 155°. Altogether, the changes were in a direction tending to minimize the recessiveness of the chin in relation to the rest of the face.

Comment: Micrognathia by itself is not sufficient to produce glossoptosis and respiratory embarrassment. If the tongue is large or even normal in size, the small recessive mandible will tend to displace the tongue distally and superiorly. It is this displacement that produces the respiratory obstruction both into the hypopharynx and into the posterior choanae. On the other hand, if the tongue is small, there will be no obstruction of the airway even in the presence of a micrognathic mandible. In this instance, the simultaneous occurrence of micrognathia and microglossia averted the respiratory difficulties commonly experienced in such instances.

The tendency to lose reflex control of the muscles of respiration and deglutition under anesthesia or sedation renders such procedures unusually hazardous in these patients because of the limited reserve. It is, therefore, important that such procedures be undertaken with full knowledge and anticipation of possible respiratory obstruction, in order that adequate emergency provisions for the establishment of an airway be available.
8.1.3 Case 3

E.C., a white boy aged 5 weeks, was referred to the outpatient clinic of the Cleft Palate Center for longitudinal growth studies. The delivery had been normal and at full term. His birth weight was 7 lb., 8 oz. (3,400 gm.). There was no history of cleft on either side of the family. No difficulty in breathing was encountered, and the infant was discharged from the hospital on the sixth day. After a brief adjustment period, the infant was readily fed by a combination of a hard nipple and by means of a premature baby bottle nipple.

Some snoring sounds were heard, especially as the infant was placed on its back and the head elevated with slight ventroflexion on the chest. The infant preferred to sleep on either side, and in these positions the snoring sounds were at a minimum. This baby

---

**Fig. 8.3 a, b.** Palatal growth changes in a child with a Pierre Robin sequence. This sequence is characterized by glossoptosis, micrognathia, and isolated cleft palate. In many cases the cleft plate which is initially wide at birth can spontaneously narrow with palatal growth. a Computer-generated tracings of the isolated cleft of the hard palate from 2 months and 10 days (0-2-10) to 6 years, 1 month, and 1 day (6-1-1). The palate was closed at 4 years, 2 months. b Superimposed tracings of each cast [on the baseline created by connecting postgingivale points (the posterior limits of the hard palate) and registered at the bisector of the line] show that the length of the cleft increases with palatal growth and narrows due to spontaneous growth at the medial border of the palatal processes. Obturators which interfere with tongue posturing within a relatively small intraoral space are contraindicated [16]
showed progressive improvement, and at the age of 5 months he weighed 15 lb., 8 oz. (7,030 gm.)

**Growth Studies:** Two sets of records are available in this case. The first was obtained at the age of 1 month 7 days, and the second at 3 months 25 days of age.

The first cast of maxilla revealed a wide parabolic cleft extending distally from the nasopalatine foramen. The widest portion of the cleft, at the level of the maxillary tuberosities, measured 16 mm. Although the second cast exhibited an increase in the length and width of the palate, there was a decrease of 1.5 mm in the width of the cleft at its widest portion. During the first examination, the tongue was observed to occupy at rest the opening into the nasal chambers provided by the cleft in the palate. This was further confirmed by examining the frontal and lateral views of the head plates. The second series of films indicated that the tongue was now postured in a more inferior position and no longer occupied the nasal cavity to the same extent previously noted. This new position of the tongue could be explained by the downward and forward growth of the mandible that had occurred in the interim.

In the first lateral film, the recessive chin, the distally and superiorly malposed tongue, and relatively restricted airway were clearly observed. Two and one-half months later, considerable growth in the mandible had occurred to improve the facial profile, alter the posture of the tongue, and increase the anteroposterior diameter of the airway. The tongue was no longer in close apposition to the posterior pharyngeal wall, and its superior margin did not extend into the nasal cavity to the degree previously observed. Coincidentally, the mother reported a diminution of the stertorous breathing that had been present.

Superimposition of the tracings of the bony structures revealed the rapid growth characteristic of this early period in life. In 2 1/2 months, that cranial vault and all parts of the face exhibited proportionate increases. Particularly encouraging was the amount and direction of growth displayed by the lower jaw. Mandibular growth was responsible not only for reducing the glossoptosis and increasing the airway, but for the improvement in the appearance of this baby’s face.

**Comment:** The problem presented by this baby was unique and different from the two previous cases of the partial obstruction of the airway. Diligent nursing care to determine the most comfortable position for breathing and feeding may be sufficient to tide such cases through their critical period. In some instances the prone positioning and orthostatic feeding suggested in the literature are most successful. Again, one is impressed by the remarkable potential for prolific growth during this period of life; a potential that is shared by the small mandible. It follows then, that every effort must be made to permit the realization of the baby’s potential for growth by providing an adequate airway, which, in turn, facilitates feeding. The clinical course to be followed is varied and depends on
the severity of the symptoms and principally upon the degree of obstruction of the airway.

### 8.2 Comment

The representative sampling of cases presented provides an answer to the questions which the study was designed to solve. It is observed from the data presented that the mandible possesses remarkable potentialities for growth in patients with the Pierre Robin syndrome. Thus, all efforts should be directed toward sustaining life in a metabolically favorable climate in order that a more physiologic airway may be established as growth proceeds. With growth, the glossoptosis is minimized and spontaneous resolution of the respiratory and feeding problems occurs. It is our opinion that tracheotomy should be resorted to promptly if respiratory embarrassment is significant, in order to achieve a sufficient airway to provide adequate oxygenation. This is undoubtedly a life-saving procedure in some patients.

On the basis of our longitudinal growth studies, certain prognostications concerning the future growth of the micrognathic mandible are permissible. In most instances, the increment in mandibular growth, as related to total facial growth, is sufficient to overcome the extreme recessiveness of the chin that is observed at birth. Since mandibular growth continues until late adolescence, it is possible to hope for an esthetically pleasing profile in adulthood. The management of the cleft palate has been in keeping with the established criteria for the treatment of palatal defects.

The lateral cephalometric film served as a valuable diagnostic tool in estimating the degree of obstruction of the airway as a result of the glossoptosis. In our experience, there was a high positive correlation between the degree of obstruction revealed in the x-ray film and the incidence and severity of the respiratory difficulties. When obstruction of the air passage was complete and the tongue was practically in contact with the posterior wall of the pharynx, tracheotomy was recommended as a life saving procedure. If the obstruction was incomplete, more conservative measures were employed. Care was taken to ascertain the most comfortable postures for breathing and feeding for the individual case, and the nurse or parent was carefully instructed in the care of the infant. Appropriate nipples were selected to minimize the energy expended by the infant in the feeding process.

In the course of these studies, we were aware of an obvious objection to placing so much reliance on these roentgenograms. Since many of these films were obtained under mild sedation, was it not possible that the posture of the mandible or of the tongue might have been altered by the sedative? Secondly, the film depicted a static view of the airway and represented only two dimensions. Did this view properly reflect the kinetic ability of the infant to manipulate the tongue and jaw hence the consistent correlation between the findings in our films and the clinical state? Moreover, when the films were repeated in the same infant without sedation, similar postures were recorded for the structures under analysis. It was important that the postures of the head in relation to the neck be kept constant. Dorsiflexion or ventroflexion of the head varied to posture of the mandible and tongue and produced changes in the configuration of the airway. To indicate alterations in the posture of the head to the neck, our tracings purposely included at least the first two cervical vertebrae.

We recognize that few institutions possess cephalometric roentgenographic equipment. Therefore, we should like to point out that an ordinary lateral film obtained by carefully positioning the infant can provide useful diagnostic data. To minimize enlargement, a target-object distance of at least 3 ft. (90 cm) is recommended. For the sake of definition and to further decrease enlargement, the object film distance should be kept at a minimum. Sjölin [15] has published interesting films to describe his experiences with a case of micrognathia. Although his films did not permit quantification of the growth changes, they were adequate for diagnostic purposes.

A number of papers in the literature claim to “stimulate” the growth of the mandible by a variety of mechanical devices or surgical procedures. For example, a special nursing bottle was designed to force the infant to protrude his jaw in order to obtain nourishment and, by this protrusion, to stimulate mandibular growth [4]. From our data, we would conclude that the nursing care enabled the infant to survive until mandibular growth was sufficient to provide a more adequate airway.

In another report, continuous traction on the mandible was maintained by circumferential wiring around the symphysis. The authors claimed growth-stimulating properties for this procedure [10]. From the findings in our series, it would seem that mandibular growth probably occurred spontaneously and not because of the stimulus provided by surgical traction.

The important and prime objective in the care of these children is to provide an airway. If possible, this should be accomplished with a minimum of trauma. Secondly, the infant’s total needs should be assessed to provide optimal conditions for somatic growth. As the potential for growth is permitted to express itself, the chin grows downward and forward away from the base of the skull. With this pattern of growth, adequate space for the tongue is provided, the airway...
enlarges, and there follows a spontaneous resolution of the symptoms. Also, there are progressive improvements in the facial appearance.

There is another dimension to the abnormal posture of the tongue, as observed in these patients, that merits discussion. Not only does the tongue block the pharyngeal processes and hence prevent their fusion. The high incidence of micrognathia in the population of clefts involving only the hard and soft palate lends support to this theory. Mandibular micrognathia is a physiological finding in early intrauterine life. If for some reason the micrognathia persists and fails to carry the tongue down and out of the nasal cavity, a cleft in the palate might result.

In early postnatal life, the tongue acts to keep the cleft palatal processes apart. As the tongue descends with mandibular growth and no longer forcefully intrudes itself into the nasal cavity, the palatal processes tend to approximate in the midline. Fusion of the palatal processes cannot occur, but the narrowing in the clefts is recorded fact.

8.3 Summary and Conclusions

The development of the accurate techniques for cephalometric roentgenography of infants has made possible a longitudinal study of the growth of the micrognathic mandible. As a result of these studies, useful diagnostic and prognostic information has been obtained to provide a rationale for the management of individual cases.

The lateral cephalometric roentgenogram is a valuable diagnostic aid in assessing the severity of the glossoptosis and its obstruction of the airway. A definite correlation exists between the degree of constriction of the airway and the severity of the clinical state. On the basis of these findings, it is possible to recommend either conservative management or tracheotomy in extreme situations, or distraction osteogenesis. Three cases, out of a larger series of similar cases, were presented to indicate the spectrum of variations to be encountered.

In all instances, it was found that where an adequate metabolic situation was provided and the infant gained weight, mandibular growth during the first few months was sufficient to provide for a natural resolution of the symptoms attending the glossoptosis.

Longitudinal records have indicated that mandibular growth is proportionally adequate to reduce the retrognathic profile and provide an esthetically harmonious facial appearance.

Based on investigations performed during the tenure of Special Research Fellowship from the National Institute of Dental Research Institutes of Health (Dr. Pruzansky, Senior Assistant Dental Surgeon [R], United States Public Health Service, National Institute of Dental Research, Department of Health, Education and Welfare).

References